Dermatomyositis as the Initial Presentation of Ovarian Cancer

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Dermatomyositis is a rare systemic inflammatory myopathy with characteristic cutaneous manifestations consisting of heliotrope erythema, Gottron's papules, poikiloderma, and periungual telangiectasia [1]. Diagnosis is based on characteristic skin lesions and additional laboratory examination results. Laboratory confirmation of dermatomyositis includes increased levels of serum muscle enzymes (creatine phosphokinase, aldolase, and lactate dehydrogenase) and electromyographic changes. Although the underlying pathogenesis remains elusive, the association of this disease with malignancies is well known. Documented data from Scandinavia shows increased rates of malignant diseases in patients with dermatomyositis, compared with the general population [2,3]. We report here a case of epithelial ovarian cancer with an initial presentation of dermatomyositis. The manifestations of dermatomyositis regressed in synchrony with effective treatment of the tumor with surgery and systemic chemotherapy.

A 60-year-old, multipara, menopausal woman, with no remarkable medical history, presented to our dermatology clinic with a complaint of intermittent facial erythema for about 2 months. It was mainly distributed on the bilateral cheeks and forehead. Progressive symmetrical muscle weakness, which led to difficulty in cooking and climbing upstairs, was also reported, together with progressive abdominal distension and increased abdominal circumference noted 10 days prior to admission. On physical examination, cutaneous lesions consisting of generalized confluent macules and violaceous erythema were found on the face, trunk, and all four limbs (Figure). Periorbital heliotrope flushing associated with some degree of edema, violaceous papules over the knuckles (Gottron's sign), and periungual erythema with telangiectasia were also noted. Skin biopsy was suggested but was refused by the patient. Abdominal distension was present with no palpable abdominal mass. On neurologic examination, the muscle power of the bilateral arm, forearm and thigh muscles had decreased to grade 4, while that of the unaffected muscles remained at grade 5. The results of laboratory examination revealed elevated levels of creatinine phosphokinase (102 IU/L). The autoimmune disease profile was normal, except for antinuclear antibodies, which were present at 1:160+, in a speckle pattern. Myopathy was implied by electromyography results, which showed numerous small-amplitude and short-duration polyphasic waves with early recruitment in the right biceps. The skin lesions and muscle weakness were only partially improved by steroid treatment.

Figure. Cutaneous lesions with generalized confluent macules and violaceous erythema on the patient's back.

Chest X-ray, mammography, and ear, nose and throat examinations were performed to screen for possible underlying malignancies, but the results were normal. Panendoscopy failed to disclose any lesions suggestive of malignancy. In light of the recent onset abdominal distension, a gynecologic consultation was requested. Pelvic examination disclosed an atrophic-sized uterus and impalpable bilateral adnexa. Transvaginal sonography showed bilateral adnexal tumors with profound ascites. Ascites cytology revealed adenocarcinoma. Tumor markers were determined as: CA-125, 1,322.9 IU/mL; and CA-153, 103.4 IU/mL. A chest-abdomen-pelvis

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computerized tomographic scan confirmed bilateral ovarian solid tumors measuring 4.2 × 3.4 cm and 3.1 × 2.3 cm. Dirty infiltrations with increased soft tissue density on the mesentery and massive ascites were also noted. The patient was transferred to the gynecologic ward owing to suggested ovarian malignancy.

The patient underwent uncomplicated exploratory laparotomy in which bilateral ovarian tumors with papillary growth and diffuse peritoneal seeding were observed. A total abdominal hysterectomy, bilateral salpingo-oophorectomy, infracolic omentectomy, and bilateral pelvic lymph node dissection were performed. Suboptimal debulking surgery was achieved, and the residual tumor measuring > 2 cm was mainly located in the perirectal area and over the visceral peritoneal surfaces. Results of the final histopathologic examination confirmed a high-grade serous adenocarcinoma of the ovaries with omental metastasis, classified as International Federation of Gynecology and Obstetrics stage IIIC ovarian cancer. She completed five courses of adjuvant chemotherapy with paclitaxel (175 mg/m^2) and carboplatin (area under the curve, 6 mg/mL · min). The sixth course of chemotherapy was not delivered because of impaired liver function. The skin lesions and muscle weakness resolved completely with the normalization of CA-125 levels. However, CA-125 rose again 5 months after the initial chemotherapy, and the dermatomyositis reappeared. Abdominopelvic magnetic resonance imaging revealed multiple enlarged lymph nodes in the bilateral iliac chains. Salvage chemotherapy with liposomal doxorubicin (40 mg/m^2) and carboplatin (area under the curve, 6 mg/mL · min) were prescribed for platinum-resistant recurrent ovarian cancer. The symptoms of dermatomyositis resolved dramatically after the third course of salvage chemotherapy, and the patient was still receiving treatment at the time of writing this report.

Dermatomyositis is a rare disease with an incidence of 0.7–1.0/100,000 in the general population [4]. Its clinical significance lies in the fact that it is probably a paraneoplastic event in some patients. A large retrospective study showed that up to 15% of dermatomyositis patients had underlying malignancies [3]. Ovarian, lung and colorectal cancers were frequently diagnosed both before and after the diagnosis of dermatomyositis, suggesting that these could be candidate cancers associated with the disease.

In a review of the literature, Cherin et al [5] reported six cases of dermatomyositis with ovarian cancer in a series of 56 dermatomyositis patients (including 45 women). The incidence of ovarian cancer in the female patients with dermatomyositis (13.3%) was much higher than that observed in the general female population (1%). Moreover, in women older than 40 years with dermatomyositis, there was a high incidence of ovarian cancer among the associated internal malignancies (6/28) [5]. Nevertheless, the early detection of ovarian cancer among these patients is not substantially higher. In a case series reported by Davis and Ahmed [6], six dermatomyositis patients had abdominal symptoms at presentation. All had undergone previous screening for ovarian cancer, but ovarian disease was not found in any of them. In the case series reported by Mordel et al [7], the diagnosis of dermatomyositis preceded that of ovarian cancer in most cases, with a mean interval of 10.9 months. Delayed diagnosis could be due to the insidious onset and slow progression of dermatomyositis and the limitations of imaging studies. Ninety-four percent of patients were diagnosed as stage III or IV, rendering their prognosis extremely poor [7]. Death from ovarian malignancies associated with dermatomyositis was 100%, and the mean survival time from diagnosis was 11 months (range, 0–28 months) in the series described by Davis and Ahmed [6]. Gynecologists should, therefore, be aware of the significance of the association between these two conditions.

Some previous studies have described cases of dermatomyositis presented after an established diagnosis of ovarian cancer [8]. However, as in the present case, the diagnosis of ovarian cancer may occur shortly after the diagnosis of dermatomyositis [9]. The risk of cancer was highest during the first year following the diagnosis of dermatomyositis, and dropped substantially thereafter. However, the risk of ovarian cancer did not return to the expected population value for up to 5 years after diagnosis of dermatomyositis [1]. A thorough physical examination, pelvic ultrasound and serum CA-125 assay should, therefore, be performed at the time of presentation and patients should be closely followed for several years.

In our patient, the manifestations of dermatomyositis regressed in synchrony with effective treatment of the tumor. Although dermatomyositis may follow a paraneoplastic course or may follow its own course, independent of tumor therapy [10], clinicians should be vigilant for the recurrence of muscle weakness and cutaneous manifestations that are associated with relapse of the malignant disease in most cases.

In summary, the link between dermatomyositis and ovarian cancer should be clearly emphasized. As ovarian cancer is the most lethal gynecologic cancer, clinicians should perform timely screening in patients with dermatomyositis to detect those with occult cancer, especially in female patients older than 40 years. Even if mortality cannot be prevented in these ovarian cancer patients, disability from myositis can be alleviated when the cancers are well managed.

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